# Pediatric Cochlear Implantation: Current Practices and State of the Art Technologies

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# Learner Outcomes

- To recognize that a cochlear implant is a tool, not a cure
- To develop an awareness of different etiologies associated with deafness and hearing loss
- To understand the cochlear implant evaluation process, recognize the importance of a team approach and thorough counseling, and appreciate that individual differences impact outcomes
- To recognize that therapy must be used in collaboration with this technology; with both, children can acquire age-appropriate spoken language OR be enabled to reach his or her full potential in the use of audition

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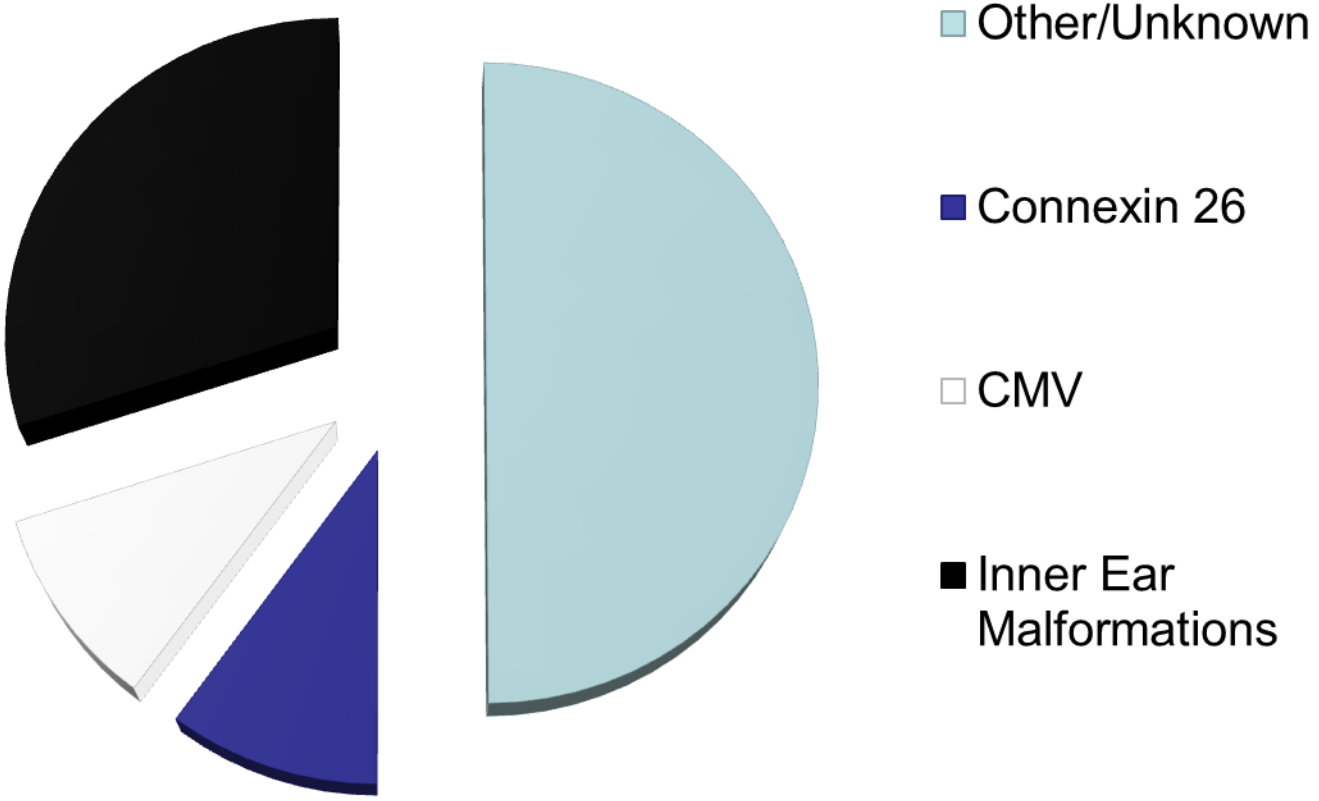
# Pediatric Hearing Loss

- Newborn Infant Hearing Screening mandated in 1999, implemented in 2001
- Annual births in North Carolina ~130,000
- 99.2% screened at birth, though not all followed up
- Estimated 3-4/1000 have hearing loss(~500)
- 1/1000 have severe to profound loss (~130)

# Etiology of Sensorineural Hearing Loss




# UNC Pediatric Audiology & CI Programs

Total 1800+ infants & children

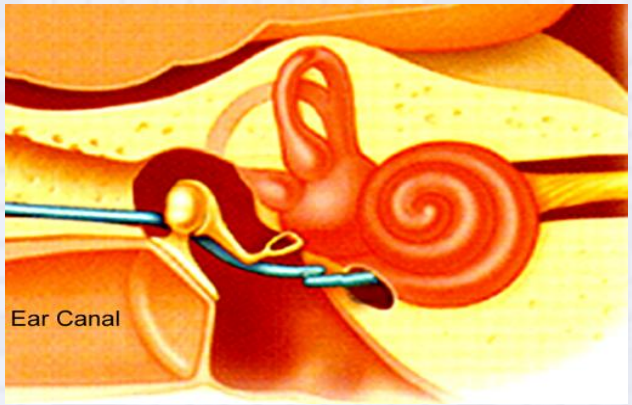
- 1000 using amplification
- 825 with cochlear implants
- 75 children using both

# What is a cochlear implant?

A cochlear implant is a prosthesis, or a tool, which is designed to detect, code and transmit the important features of sound into electrical signals that are delivered to the cochlea, the end organ of hearing.

A cochlear implant allows a deaf person to detect sound; experience with sound makes it meaningful for communication.

# A Cochlear Implant Is Different from a Hearing Aid



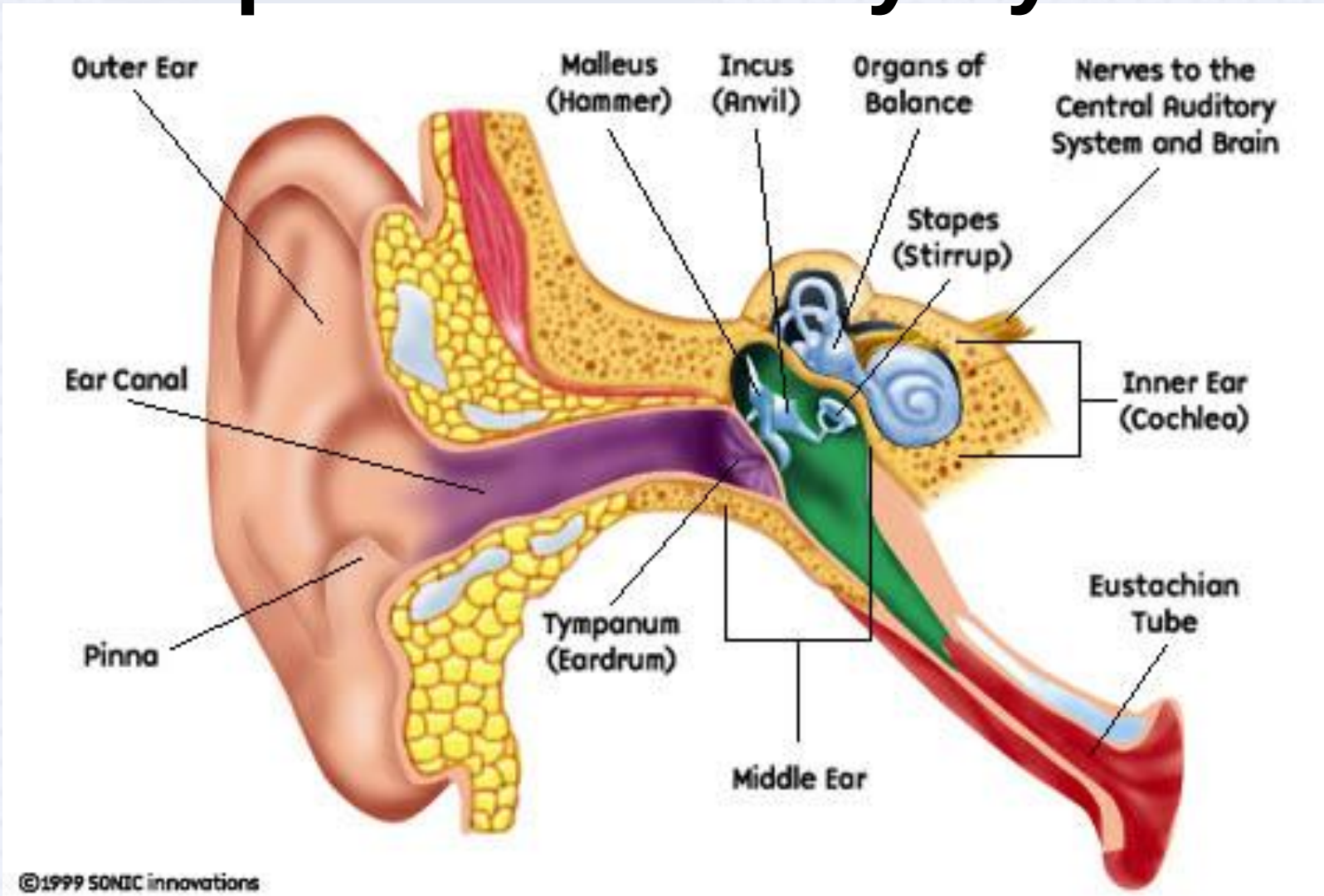

**Hearing Aids:**

- acoustically amplify sound
- rely on the responsiveness of surviving hair cells

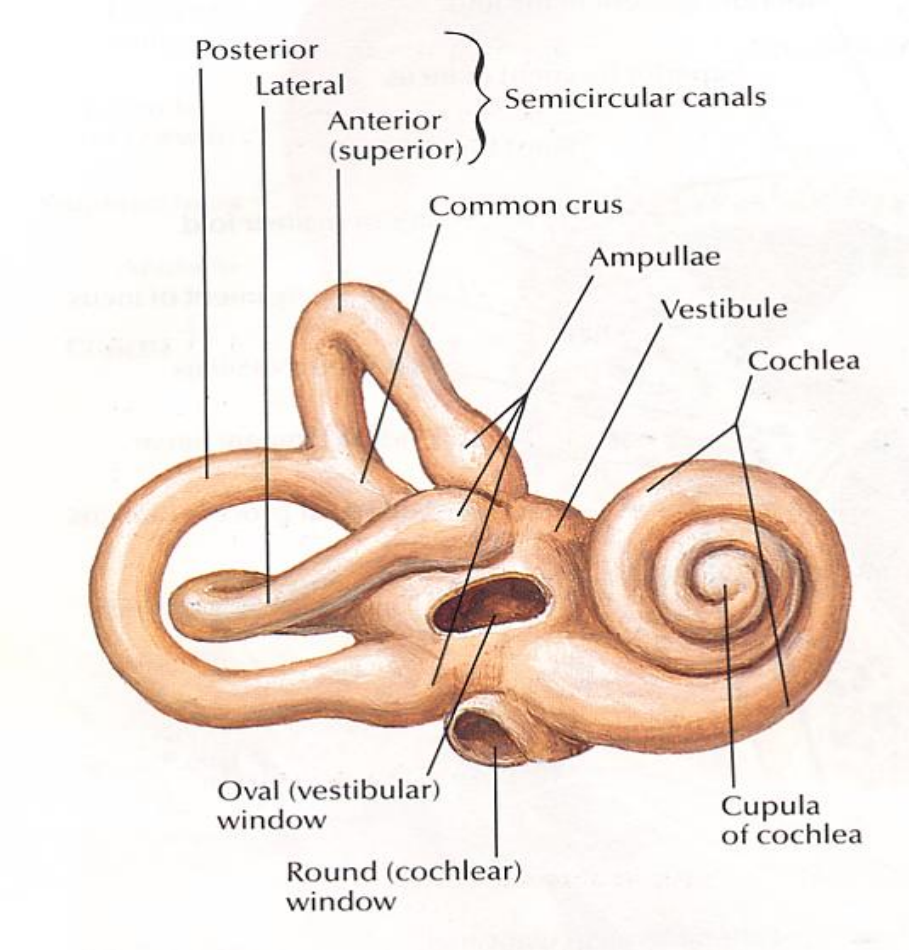
**Cochlear Implants:**

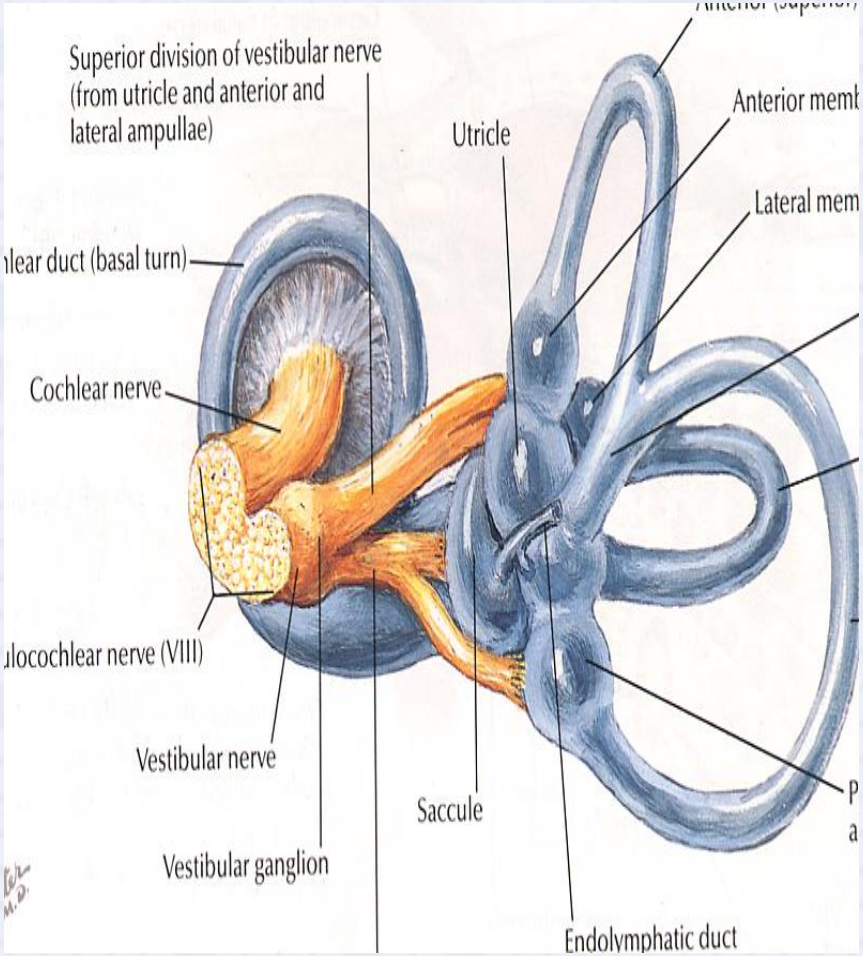
- bypass damaged hair cells and stimulate the nerve directly
- Convert the acoustic input signal into an electrical pattern recognizable by the nerve in order to yield improved speech clarity and allow speech recognition

# Peripheral Auditory System



# Anatomy of the Inner Ear



# Central Auditory Pathway

- Afferent pathways: carry sensory information from hair cells in the cochlea via the auditory brainstem to the primary auditory cortex at level of temporal lobe
- Efferent pathways:  descend from the brainstem to the cochlea
- Tonotopic organization pervasive

# Parts of the Cochlear Implant

External

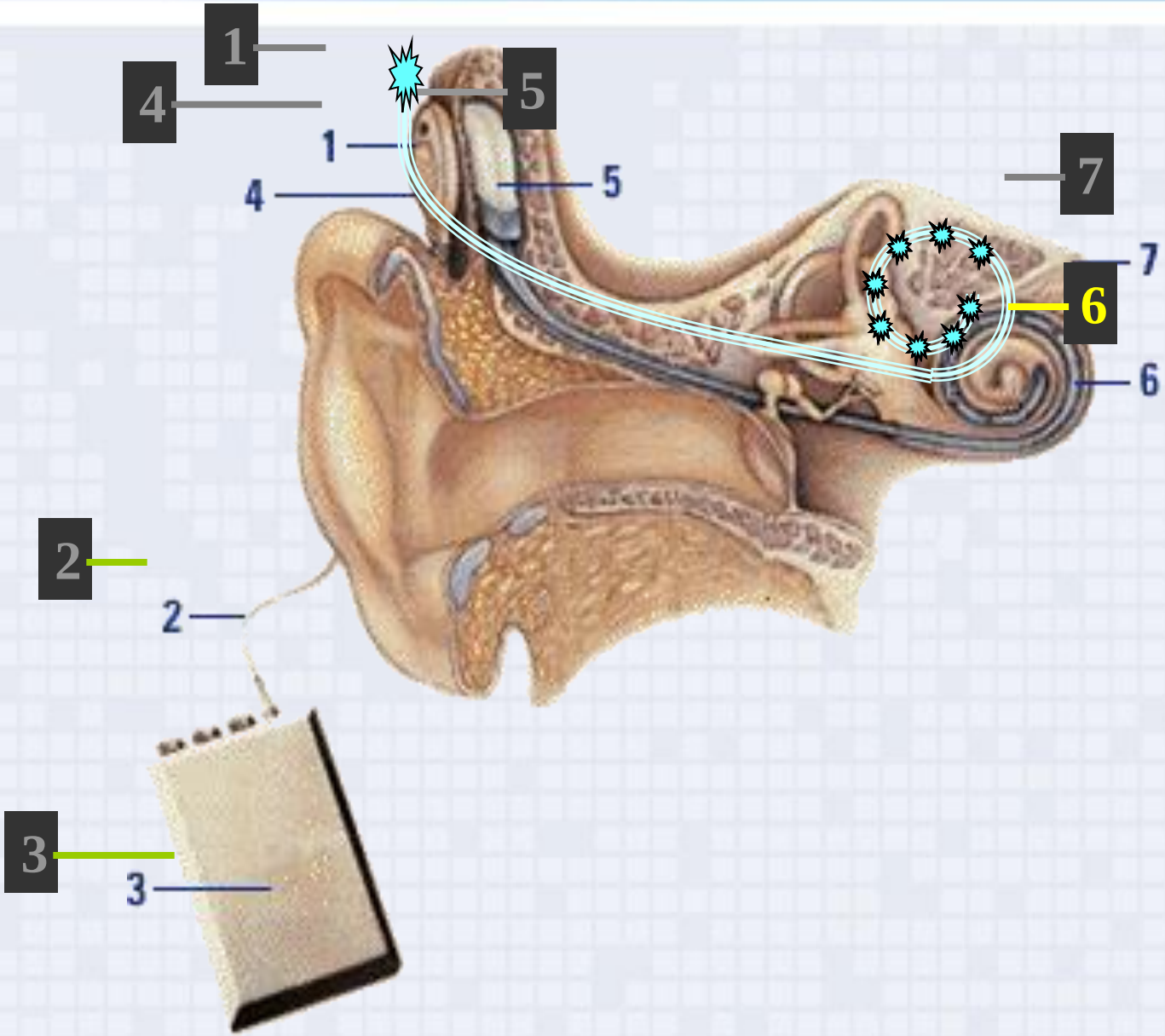
- Microphone
- Cable
- Speech processor
- Transmitting coil

Internal

- Electrode array
- Receiver/stimulator
- Magnet

# How does a cochlear implant work?

- The microphone captures sound.
- The processor converts that sound to a digital signal.
- That signal is sent through the cable to the coil.
- The signal is sent across the skin to the implant where it is converted to electrical energy.
- That energy is sent to the electrode array within the cochlea where it stimulates the hearing nerve.
- This stimulation is perceived as sound.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# Cochlear Implant Systems

- Cochlear Corporation
  - www.cochlear.com
- Advanced Bionics
  - www.advancedbionics.com
- MED-EL
  - www.medel.com

# COCHLEAR

## System 5 Processor with Remote Assistant


# Med El
# Opus 2 Processor with Fine Tuner


# ADVANCED BIONICS Harmony, PSP and Neptune Processors

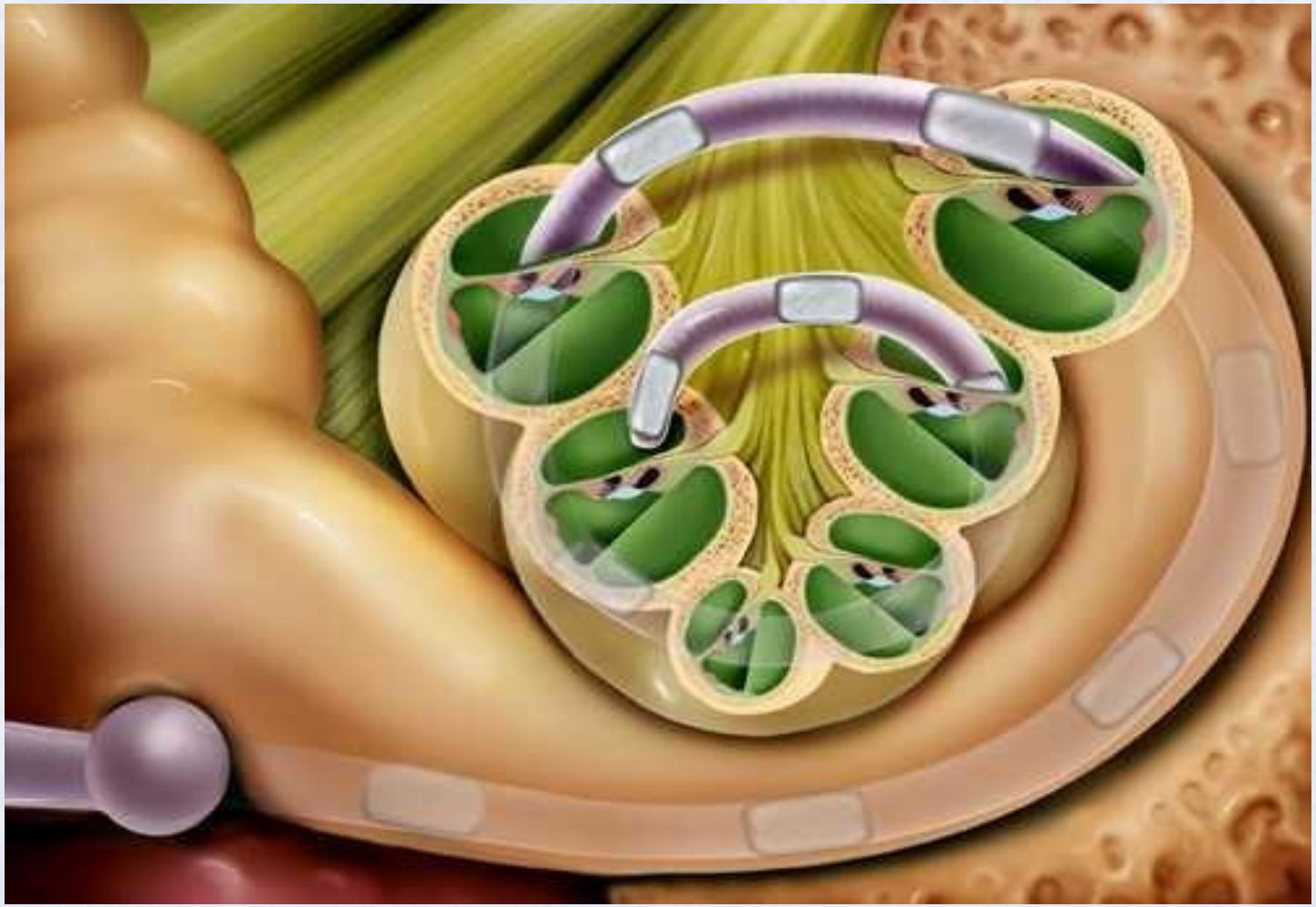


# Internal Devices

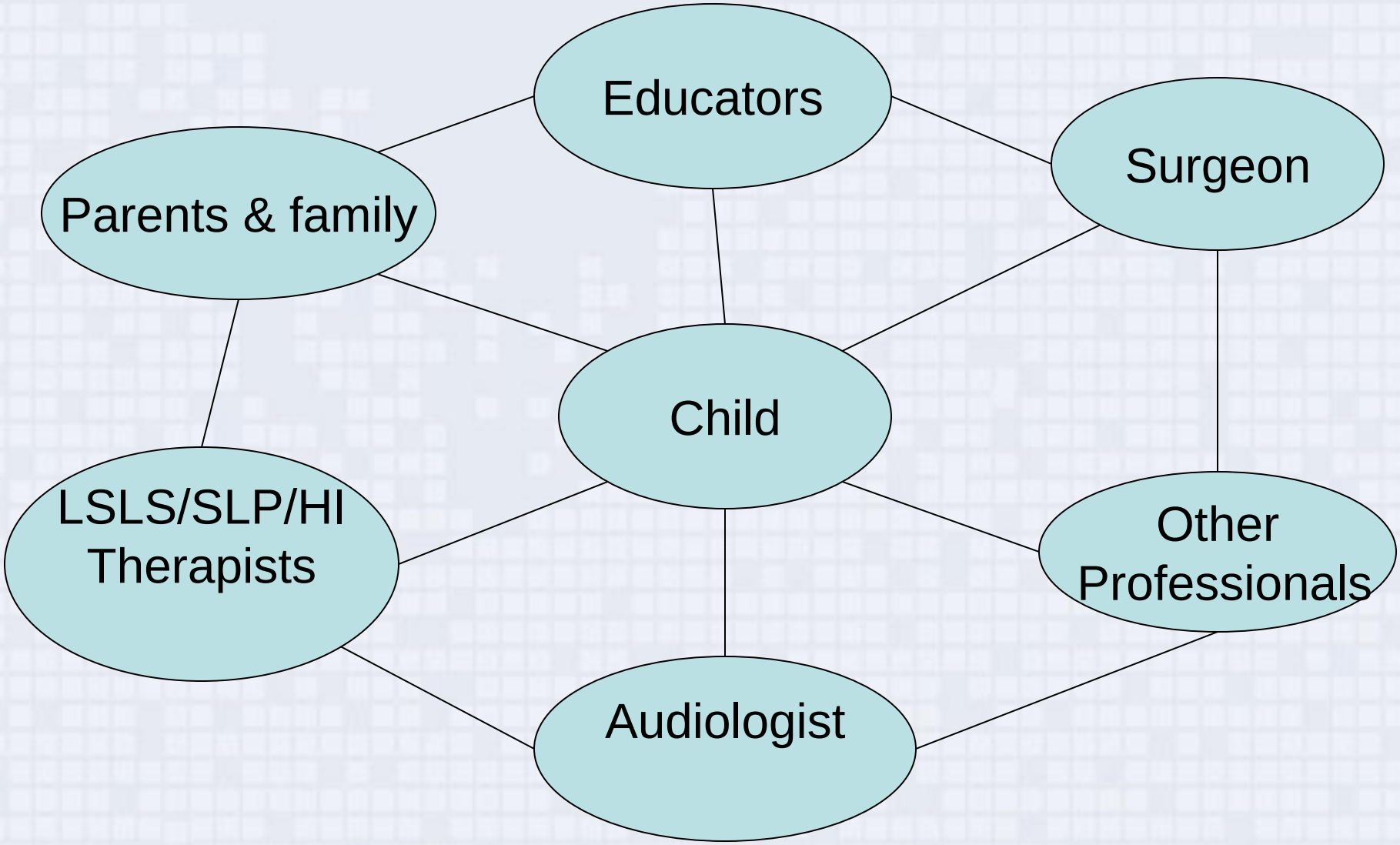
Advanced Bionics 90K


Cochlear Freedom

Med El Concert



# Pediatric Team Approach to Cochlear Implantation



# Candidacy

- Cochlear implants were once only provided to children who were profoundly deaf and had no other medical conditions
- Candidacy criteria have not changed under FDA guidelines since 2000; however, we have learned the potential benefits exist for many children outside these guidelines
- Cochlear implants can increase a child's quality of life
- The goal for every child is to achieve the level of benefit to reach his or her cognitive potential

# Evaluation for Candidacy

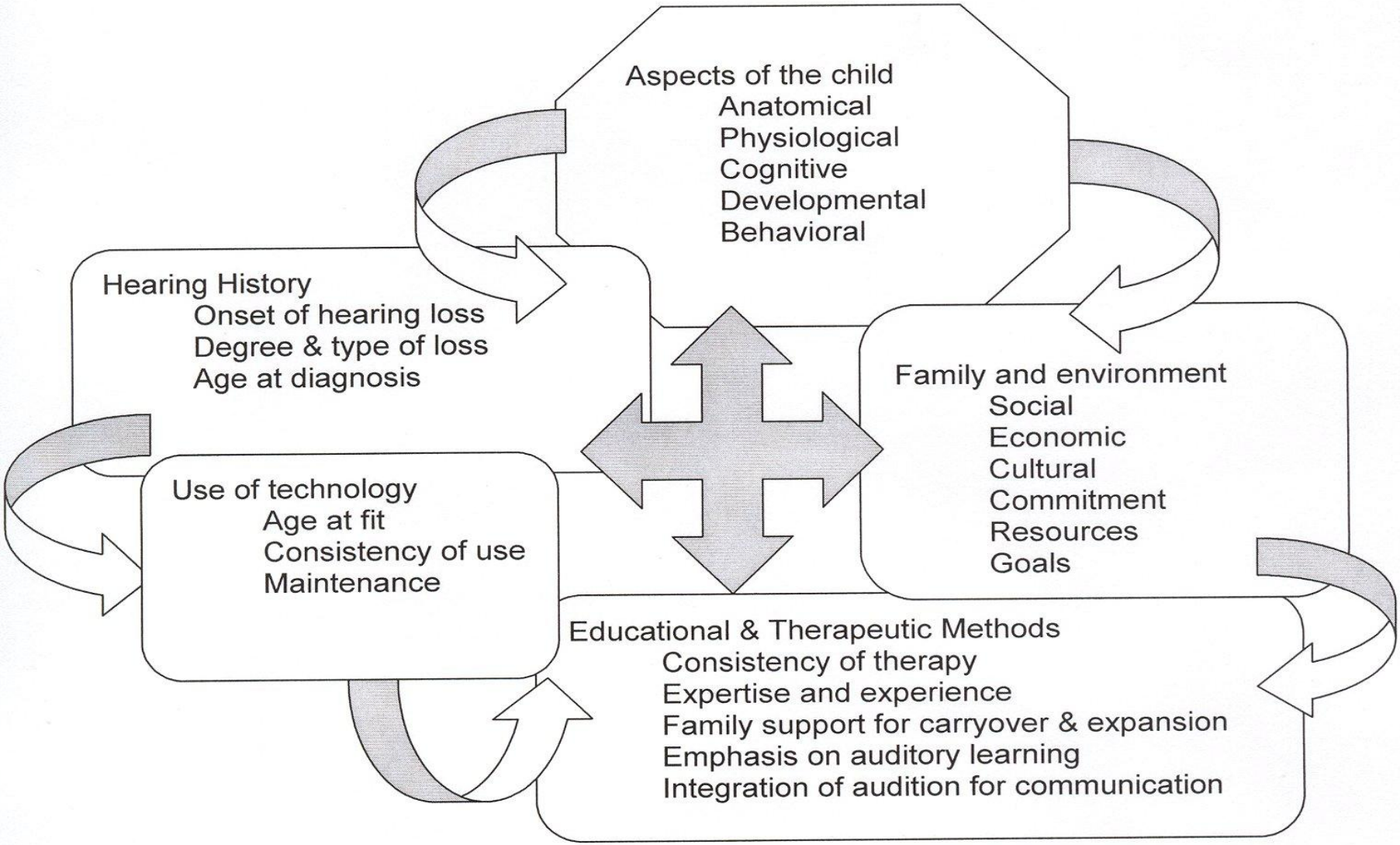
- The evaluation process can occur over several appointments, spanning up to several months
- Purpose of process is for gathering and sharing of information pertinent to candidacy and prognosis
- Includes recommendations and evaluations by numerous professionals
- Ultimate purpose:  to inform and empower the patient and/or family

# Current Candidacy Criteria: Children

- Severe to profound deafness
- Less than 30% on speech perception measures
- Lack of progress in auditory skills development
- 12 months of age
  - Exceptions
- Medically fit for surgery
- Parental support

# Additional Considerations

- Medical
  - CT/MRI Scan to visualize anatomy
  - Fit to undergo general anesthesia & surgery
  - Determine cause of hearing loss, if possible
  - Active middle ear disease
  - Other Diagnoses


Aspects of the child
Anatomical
Physiological
Cognitive
Developmental
Behavioral
Hearing History
Onset of hearing loss
Degree & type of loss
Age at diagnosis
Family and environment
Social
Economic
Cultural
Commitment
Resources
Goals
Use of technology
Age at fit
Consistency of use
Maintenance
Educational & Therapeutic Methods
Consistency of therapy
Expertise and experience
Family support for carryover & expansion
Emphasis on auditory learning
Integration of audition for communication

# Factors that Delay Implantation

- Auditory
  - Delay in diagnosis
  - Fluctuating hearing loss
  - Unreliable/conflicting test results
  - ANSD
  - Mismanagement of amplification fitting
    - Need Pediatric Audiologist with experience and proven protocols for hearing aid fitting

# Factors that Delay Implantation

- Medical
  - Anatomy not typical
    - Severe cochlear malformation
    - Cochlear Nerve Deficiency
    - Other medical diagnoses (CP, Autism
- Social challenges
  - Parental issues
  - Lack of educational/therapeutic support

# Surgery

- Similar risks to other ear surgeries
- General anesthetic
- 1.5 – 4 hours
- Ambulatory vs. Inpatient
- Full recovery typically in a few days

# Meningitis Vaccinations

- **Pneumococcal Vaccinations recommended for all patients**

– **PCV 7 (Prevnar 7) series of 4 2-15 months)**
– **PCV-23**
– **PCV-13 (Prevnar -13)**

# Initial stimulation

- Typically 3-4 weeks after surgery
- Age appropriate tasks to obtain behavioral response
- Objective responses:  NRT/NRI/ART
- Priorities are comfort and then sound awareness
- Counseling for expectations and use

# Expectations for Progress

- Goal for initial stimulation is comfort and acceptance
- Parents are taught to care for and manage the hardware
- Some children don't overtly respond the first day
- This is a work in progress.  Each child progresses at different rate
- Encourage parents and therapists to share information with CI team

# Follow-up appointments

**For children…**

- **6-8 visits the first year**
- **2-3 visits annually until older**
- **Annual visits as needed for maintenance and**

**evaluation to ensure child is making all potential progress**

# Expectations for Progress

- Encourage immediate full-time use
- First attempt at audiogram at 5 weeks post initial stimulation
- By three months, want to be in the speech banana or see changes in the child's vocalizations that suggest that they are hearing conversational level speech.
- By six months, want a stable detection audiogram at 30dBHL

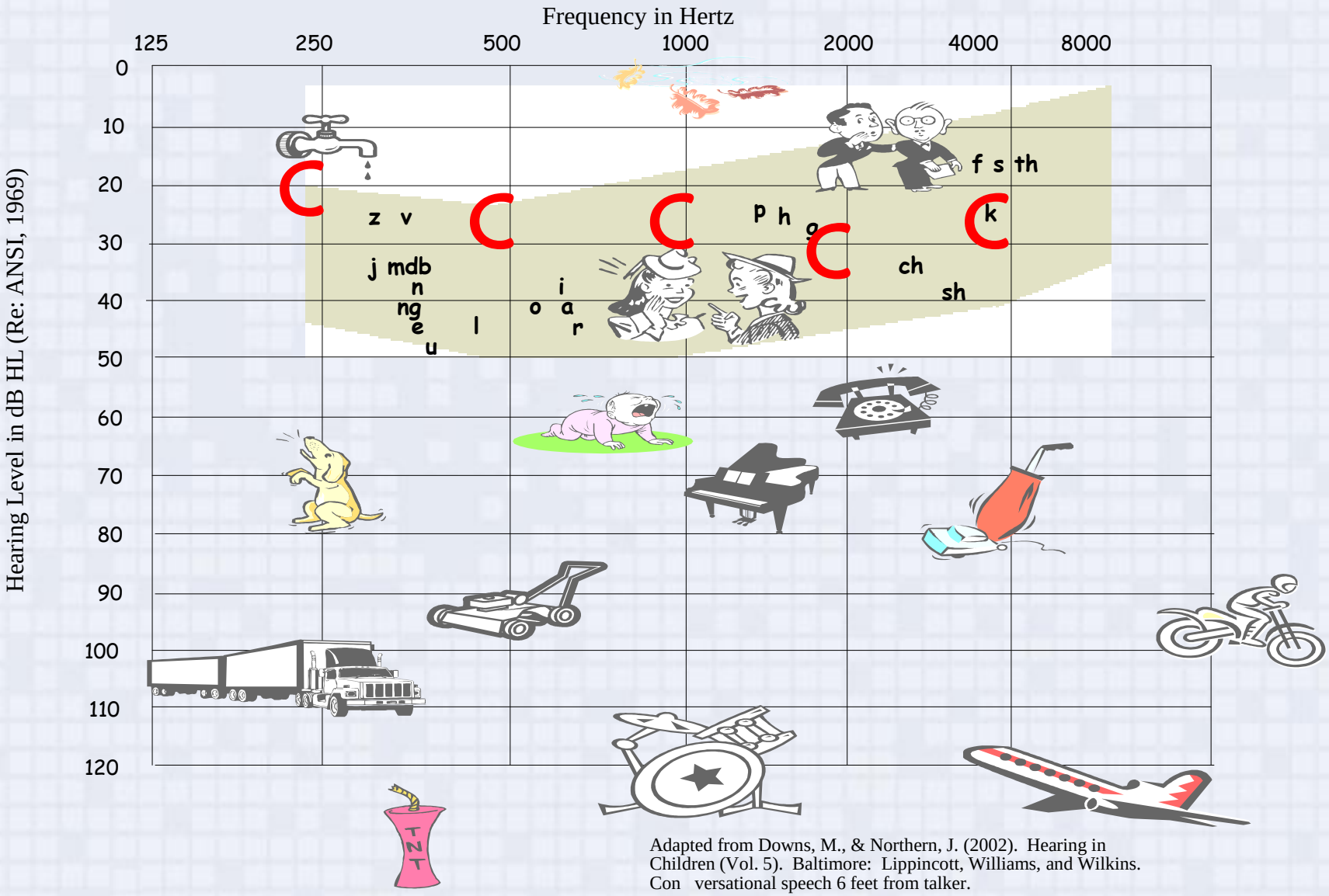
# Expectations for Progress

- By 6 months:
  - Identify several Learning to Listen Sounds
  - Use a variety of vowels
  - Identify songs through audition
  - Identify early stereotypical phrases
- See Auditory Learning Guide
- At least a year's progress in a year's time

# Speech Detection Results

- Aided "CI" audiograms all tend to look similar
  - Detection of warble tones across test frequencies from 250 - 6000 Hz in the 20 - 45 dB HL range
- There is ample evidence in the literature that cochlear implant use improves
  - Speech perception
  - Speech production
  - Language
  - Educational outcomes/ Vocational options
  - Quality of life

# Familiar Sounds Audiogram



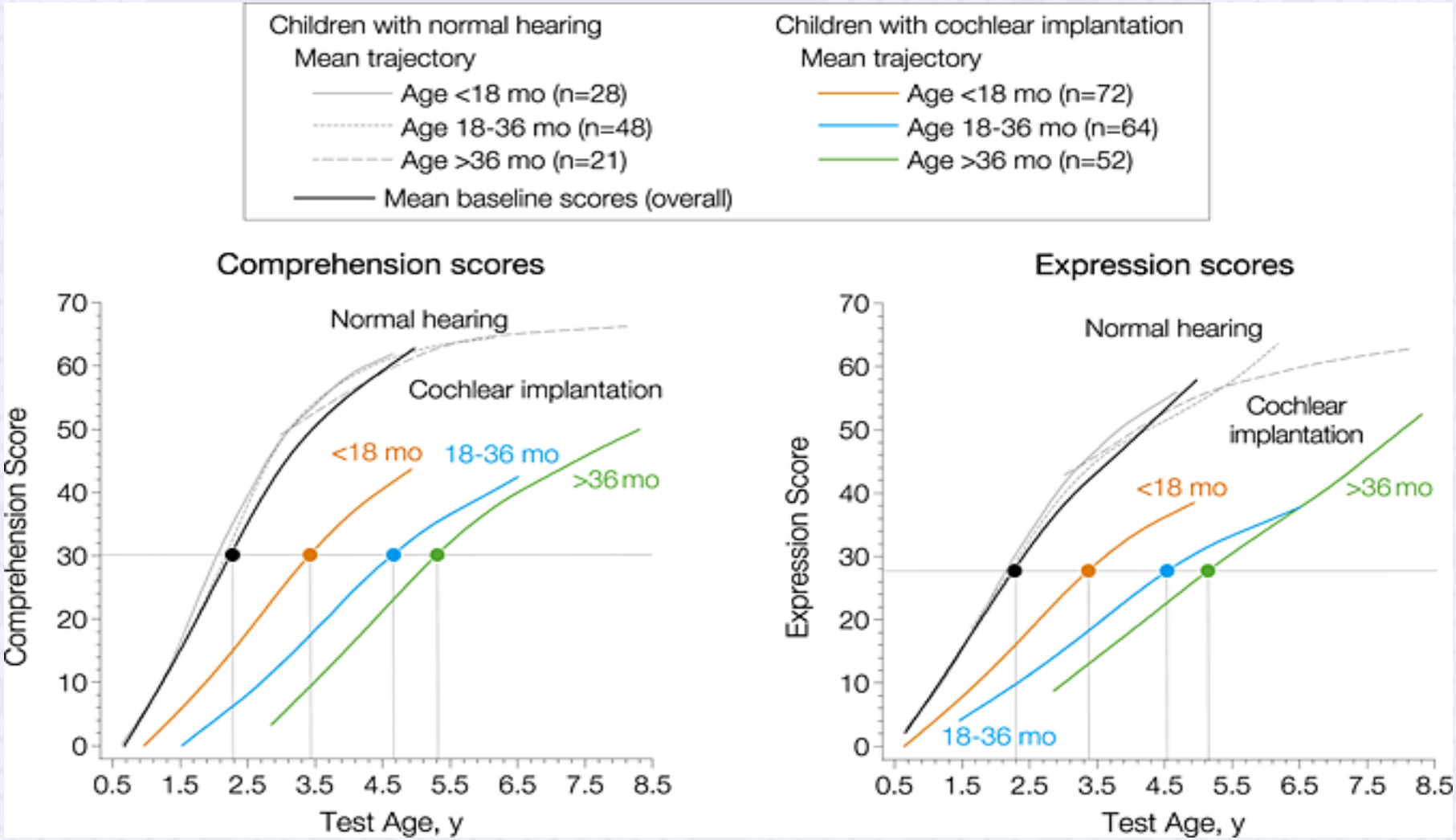

Adapted from Downs, M., & Northern, J. (2002). Hearing in Children (Vol. 5). Baltimore: Lippincott, Williams, and Wilkins. Con versational speech 6 feet from talker.

# Success with a cochlear implant

- Each child has a unique result and definitions of success vary by child
- Family commitment-regular use and device maintenance
- Early implantation
- Educational placement which provides a spoken language environment
- Teachers and therapists trained in a Listening and Spoken Language approach that integrates speech, language & audition
- Team approach
- Aggressive and frequent programming adjustments

- **When well fit and consistently used (CIs) audition may be a child's strongest modality.**
- **If implanted early, many children will be able to "catch up" from effects of auditory deprivation and develop and maintain age-appropriate spoken communication.**
- **The aim of the CI may not be to learn to talk but may be to increase auditory and communication skills, increase number of social interactions, and achieve greater connection with the environment.**

# Reynell Developmental Language Scores




**Niparko, J. K. et al. JAMA 2010;303:1498-1506.**

# Early Intervention is the Key

- With early identification through mandated newborn screening, children with severe-profound hearing losses can begin auditory-based intervention before the critical 6 month milestone.
- With today's technology, children with profound hearing loss can have full access to the speech spectrum.
- With a therapist trained in teaching spoken language through listening, early identification and technological advances, deaf children can develop age appropriate spoken language.